



Medical Coverage Policy

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Treatment of Cutaneous and/or Deep Tissue Hemangioma, Port Wine Stain and Other Vascular Lesions

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Related Coverage Resources

INSTRUCTIONS FOR USE

The following Coverage Policy applies to health benefit plans administered by Cigna Companies. Certain Cigna Companies and/or lines of business only provide utilization review services to clients and do not make coverage determinations. References to standard benefit plan language and coverage determinations do not apply to those clients. Coverage Policies are intended to provide guidance in interpreting certain standard benefit plans administered by Cigna Companies. Please note, the terms of a customer's particular benefit plan document [Group Service Agreement, Evidence of Coverage, Certificate of Coverage, Summary Plan Description (SPD) or similar plan document] may differ significantly from the standard benefit plans upon which these Coverage Policies are based. For example, a customer's benefit plan document may contain a specific exclusion related to a topic addressed in a Coverage Policy. In the event of a conflict, a customer's benefit plan document always supersedes the information in the Coverage Policies. In the absence of a controlling federal or state coverage mandate, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of 1) the terms of the applicable benefit plan document in effect on the date of service; 2) any applicable laws/regulations; 3) any relevant collateral source materials including Coverage Policies and; 4) the specific facts of the particular situation. Each coverage request should be reviewed on its own merits. Medical directors are expected to exercise clinical judgment and have discretion in making individual coverage determinations. Coverage Policies relate exclusively to the administration of health benefit plans. Coverage Policies are not recommendations for treatment and should never be used as treatment guidelines. In certain markets, delegated vendor guidelines may be used to support medical necessity and other coverage determinations.

Overview

This Coverage Policy addresses the treatment of cutaneous and/or deep tissue hemangioma, port wine stain and other vascular lesions.

Coverage Policy

Coverage for the treatment of a cutaneous hemangioma, port wine stain, or other vascular lesion varies across plans. Refer to the customer's benefit plan document for coverage details.

If coverage is available for treatment of a cutaneous and/or deep tissue hemangioma, port wine stain, or other vascular lesion, the following conditions of coverage apply.

Laser destruction (CPT codes 17106, 17107, 17108) of cutaneous vascular lesions is considered medically necessary for ANY of the following:

- port wine stain on the face and/or neck
- port wine stain on the trunk or extremities associated with recurrent bleeding or painful nodules
- cutaneous and/or deep hemangioma or other vascular malformation (e.g., venous, arteriovenous, lymphatic) and EITHER of the following indications:
 - the lesion is affecting a vital structure (e.g., nose, eyes, ears, lips, or larynx)
 - the lesion results in **ANY** of the following:
 - bleeding
 - eating difficulty
 - pain
 - repeated infection
 - swallowing difficulty
 - ulceration

Vascular embolization/occlusion (CPT codes 61626, 37241, 37242) of cutaneous and/or deep tissue hemangioma or other vascular malformation (e.g., venous, arteriovenous, lymphatic) is considered medically necessary for EITHER of the following indications:

- the lesion is affecting a vital structure (e.g., nose, eyes, ears, lips, or larynx)
- the lesion results in **ANY** of the following:
 - bleeding
 - eating difficulty
 - high output heart failure
 - pain
 - repeated infection
 - swallowing difficulty
 - ulceration

Laser destruction (CPT codes 17106, 17107, 17108) of cutaneous vascular lesions for ANY other indication is considered cosmetic and not medically necessary.

INPATIENT HOSPITALIZATION

Inpatient hospitalization of an infant for administration of oral propranolol for the treatment of cutaneous and/or deep tissue hemangioma is considered medically necessary when the lesion is ulcerated or affecting a vital structure (e.g., nose, eyes, ears, lips, larynx) and the infant is either of the following:

- age 8 weeks or less
- age 9 weeks to 12 months with ANY of the following:
 - lack of social support for home monitoring
 - presence of comorbid cardiovascular or respiratory conditions
 - presence of a comorbid condition affecting glucose levels

General Background

Vascular lesions may be classified into two main categories: vascular tumors and vascular malformations. Vascular tumors are characterized by vascular endothelial cell hyperplasia and spontaneous involution. The most common vascular tumors are hemangiomas.

Vascular malformations are abnormalities in blood vessel formation; these lesions do not regress and slowly enlarge. The name of the malformation reflects the blood vessel forming the lesion: capillary, venous, arterial or lymphatic. A common capillary malformation, the port wine stain, is characterized by flattened endothelial cells with normal turnover. Venous malformations give a bluish color to the area under the involved skin or mucosa. Arterial malformations are rare, are often referred to as arteriovenous malformations and are direct connections of arteries to veins. Lymphatic malformations can involve either large (cystic hygroma) or small vessels (lymphangioma circumscriptum). A fibro-adipose vascular anomaly (FAVA) is an anomaly in which fibrous fatty

tissue replaces muscle tissue, and becomes intertwined with veins and/or lymphatic vessels. Vascular malformations can also consist of combinations, such as with Klippel-Trenaunay Syndrome or Sturge-Weber Syndrome.

Vascular lesions may result in permanent disfigurement with the main goal of treatment aimed at improving cosmesis. However, some vascular lesions interfere with functioning of vital structures or result in symptoms, such as pain, ulceration and bleeding. In general, treatments are dependent on the type and severity of the lesion but may include any of the following procedures, which may be performed alone or in combination:

- injectable medications (e.g., corticosteroids)
- laser therapy
- sclerosant therapy
- embolization
- surgical debulking, with or without compression garments
- radiotherapy

Cutaneous/Deep Tissue Hemangiomas

Cutaneous hemangiomas occur in approximately 1 out of 10 children; these lesions are characterized by rapid proliferation in early infancy and slow involution that may occur over several years. The mechanisms that control involution are not well understood. Some hemangiomas are present at birth as precursor lesions; rarely are they fully formed tumors at birth. More commonly, lesions become evident after birth, usually within two to four weeks. Hemangiomas frequently occur on the face and neck area but may also be located on areas such as the trunk and/or internal organ structures. Most hemangiomas do not require medical intervention, although a small number cause functional complications or disfigurement. Permanent disfigurement is more likely if lesions are present on the face; the nose, lip, and forehead are most vulnerable (Conlon and Drolet, 2004).

Several types of congenital hemangiomas have been described in the literature and include those that are rapidly involuting, and noninvoluting. Kasabach-Merritt syndrome is a complication of rapidly enlarging vascular lesions (hemangioma) and is characterized by hemolytic anemia, thrombocytopenia and coagulopathy. While these lesions are not hemangiomas of infancy, they result from a more aggressive proliferative vascular tumor that results in decreased platelets and other bleeding problems. Although the syndrome is rare it requires aggressive treatment and is often associated with a high mortality rate.

For a majority of hemangiomas no intervention is needed and lesions regress spontaneously. Some lesions result in untoward cosmetic changes that have no clinical significance. Although infantile hemangiomas involute, involution may be incomplete with resulting permanent skin changes that may be life altering, particularly for lesions that are thick (Krowchuk, et al., 2019). Complications resulting from hemangiomas have been reported and are often related to the site of occurrence, with approximately 10-20% of cases requiring treatment due to complications (Satterfield, et al., 2019). The most frequent complication associated with hemangiomas is ulceration which is often present during the proliferative phase. Periorbital hemangiomas may cause amblyopia, impaired vision and astigmatism and should be considered when a hemangioma involves the eyelids or periorbital tissue. Approximately 43-60% of individuals with periorbital lesions can develop amblyopia (Al Dhaybi, et al., 2011, Leaute-Labreze, et al., 2011). Lesions located on the ear may result in auditory impairment and secondary speech delay. Subglottic hemangiomas may cause hoarseness and stridor leading to respiratory impairment and are associated with at least 50% mortality if untreated (Peridis, et al., 2011). Many patients with subglottic hemangiomas also have cutaneous hemangiomas involving the lips, chin, and mandible. Hemangiomas may also be located on the cervicofacial area and lumbosacral spine. Pedunculated hemangiomas may be at risk for bleeding and irritation and have been associated with permanent cosmetic skin changes after involution such as fibrofatty tissue and excessive scarring. Although infrequent, larger some hemangioma lesions may result in high-output heart failure due to high flow.

The main goals of treatment include preventing permanent disfigurement and minimizing psychosocial distress, preventing functional complications, and treating ulceration. Several modalities have been proven effective to treat hemangiomas and include the administration of steroid medications as the mainstay of treatment (e.g., topical, intralesional and systemic), pulsed dye laser therapy, and interferon. Corticosteroids have been associated with significant adverse events such as Cushing's syndrome, hypertension, immunosuppression,

hyperglycemia and adrenal suppression (Hogeling, et al., 2011). The pulsed-dye laser has been proven effective for the treatment of superficial hemangiomas, the superficial component of mixed hemangiomas and ulcerated hemangiomas. Efficacy is however limited by the depth of laser penetration. Several treatments may be necessary, and treatments have been associated with some risk of scarring (Rudolph, 2003). Other, less common treatments include: cryotherapy, other forms of laser surgery, embolization, and use of chemotherapeutic agents, such as vincristine and cyclophosphamide. Embolization may be required to control arteriovenous shunting that may be present in large congenital hemangiomas, (e.g., >7 cm) to prevent cardiac overload and subsequent high-output cardiac failure (Ricci, et al 2017). Other forms of laser surgery have included the argon laser for hemangiomas, the Nd:YAG (neodymium: yttrium-aluminum-garnet) laser for deeper lesions, and carbon dioxide laser for lesions such as subglottic hemangiomas. Some of these devices have been associated with significant scarring. Surgical excision may be recommended for hemangioma lesions that are sharply demarcated and pedunculated, are ulcerated and bleeding, have not responded to other modalities of treatment, and those that threaten function (Rudolph, 2003).

Oral Propranolol: Oral propranolol has been utilized as a treatment for infantile hemangioma, and is considered first-line treatment for infantile hemangiomas at risk for complications. Infantile hemangiomas occur in approximately 4% to 5% of newborns (Chamli, Litaïem, 2019; Wildgruber, et al., 2019). This type of treatment is aimed primarily at lesions that interfere or have potential to interfere with vital function and/or are life-threatening (Satterfield, et al., 2019; Krowchuk, et al., 2019; Drolet, et al., 2013). In some cases treatment may be recommended to improve cosmesis when there is risk of permanent disfigurement (Krowchuk, et al., 2019; Drolet, 2013).

Propranolol, a non-selective beta-blocker, exerts a vasoconstricting effect which may result in a change in color, reduction of lesion volume, softening and regression of the lesion. Induction of apoptosis is also a possible mechanism of action for reducing hemangioma lesions. Initiation of therapy may be performed in a hospital setting as either inpatient or outpatient depending on the resources available for safe monitoring. Although specific dosing, age for initiation of therapy, duration of treatment, and expected clinical outcomes are not firmly established, treatment protocols have been published.

Evidence evaluating the use of propranolol as a treatment for infantile hemangioma is in the form of case reports, retrospective or prospective case series, and uncontrolled comparative trials involving small populations. Published randomized and/ or controlled trials are limited; however studies are currently being conducted through the U.S. National Institute of Health to further evaluate safety and efficacy. Published data indicate the type of hemangioma lesion most often treated is a clinically compromising lesion, such as orbital or airway lesion. Age for initiation of therapy has ranged from one month to five years although most subjects were less than 12 months of age. Reported efficacy is variable but tends to be higher when administered during infancy and the proliferative phase of involution, although regression of lesions has been documented when administered during the involution phase. Duration of therapy within these trials ranged from one to 12 months with six months being the average. Follow-up evaluation of clinical outcomes varied as well, ranging from immediately following initial treatment to 18 months post-treatment.

In 2019 the American Academy of Pediatrics (AAP) published clinical guidelines regarding management of infantile hemangiomas (IHs) (Krowchuk, et al, 2019). Within this guidelines high risk IHs are defined as having evidence of or potential for those that result in life-threatening complications, functional impairment, ulceration, structural anomalies (eg, in PHACE syndrome or LUMBAR syndrome) or permanent disfigurement (X; strong recommendation). Oral propranolol is considered as the first-line agent for IHs requiring systemic treatment (A; strong recommendation). Specific recommendations regarding pharmacotherapy are as follows:

- Dose propranolol between 2 and 3 mg/kg per day unless there are comorbidities (e.g., PHACE syndrome) or adverse effects (e.g., sleep disturbance) that necessitate a lower dose (A; moderate recommendation)
- Counsel that propranolol be administered with or after feeding and that doses be held at times of diminished oral intake or vomiting to reduce the risk of hypoglycemia (X; strong recommendation)
- Evaluate patients for and educate caregivers about potential adverse effects of propranolol, including sleep disturbances, bronchial irritation, and clinically symptomatic bradycardia and hypotension (X; strong recommendation)

- May prescribe oral prednisolone or prednisone to treat IHs if there are contraindications or an inadequate response to oral propranolol (B; moderate recommendation)
- May recommend intralesional injection of triamcinolone and/or betamethasone to treat focal, bulky IHs during proliferation or in certain critical anatomic locations (e.g., the lip) (B; moderate recommendation)
- May prescribe topical timolol maleate as a therapy for thin and/or superficial IHs (B; moderate recommendation)

Surgical management and laser therapy may be recommended as treatment options in managing selected IHs (C; moderate recommendation).

AAP grading of recommendations and evidence for the above interventions is as follows:

- Level A- well-designed and conducted clinical trials, meta-analyses on applicable populations
 - Level B- trials or diagnostic studies with minor limitations; consistent findings from multiple observational studies
 - Level C- single or few observational studies or multiple studies with inconsistent findings or major limitations
 - Level D- expert opinion, case reports, and reasoning first principles
 - Level X- exceptional situation in which validating studies cannot be performed and there is a clear preponderance of benefit or harm.
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- Strong recommendation- action is favored because anticipated benefits clearly exceed harms (or vice versa) and quality of evidence is excellent or unattainable.
 - Moderate Recommendation-action is favored because anticipated benefits clearly exceed harms (or vice versa) and the quality of evidence is good but not excellent (or is unattainable)
 - Weak Recommendation (based on low quality evidence) - action is favored because anticipated benefits clearly exceed harms (or vice versa), but quality of evidence is weak
 - Weak Recommendation (based on balance of benefit and harms) - aggregate database shows evidence of both benefit and harm that appears to be similar in magnitude for any available courses of action.

Results of early clinical trials demonstrate clinical effectiveness as early as 24 hours following administration with reduction of volume, change in color from red to purple, and softening of the lesion (Leaute-Labreze, et al., 2011; Manunza, et al., 2010; Sans, et al., 2009). Complete regression in as little as two months following treatment has been reported (Sans, et al., 2009) and a majority of the published evidence since this early trials demonstrate positive response rates with partial to complete regression of lesions and minor side effects (Zhang, et al., 2017; El Hachem, et al., 2017; Léauté-Labrèze, et al., 2015; Léauté-Labrèze, et al., 2013; Luo, et al., 2014; Sharma, et al., 2013; Vassallo, et al., 2013; Hermans, et al., 2012; Ming-ming, et al., 2012; El-Essawy, et al., 2011; Spiteri Cornish and Reddy, 2011; Thoumazet, et al., 2011; Hogeling, et al., 2011; Price, et al., 2011; Schupp, et al., 2011; Leaute-Labreze, et al., 2011; Al Dhaybi, et al., 2011; Manunza, et al., 2010; Buckmiller, et al., 2010; Sans, et al., 2009). In addition to regression of the lesions, improved clinical outcomes such as decrease in astigmatism, improved amblyopia (Vassallo, et al., 2013) and decreased airway obstruction (Hermans, et al., 2012) have been reported.

Peridis et al. (2011) published a meta-analysis on the effectiveness of propranolol for the treatment of infantile airway hemangioma and compared propranolol with other therapies. Included in this review was a statistical analysis of variables using an odds ratio and sensitivity analysis. Thirteen studies met inclusion criteria involving 36 subjects in total. The authors reported propranolol was an effective treatment for resolution of lesions ($P < 0.00001$), and was significantly more effective than steroids ($P = 0.0002$), CO₂ laser ($P = 0.0005$), and vincristine ($P = 0.01$). It was noted propranolol decreased airway stenosis after one week of therapy from an average of 77.57% to 38.3% and after 4 weeks to an average of 24.6%. Only one child developed complications related to propranolol which was bronchoconstriction during the first week of therapy. Although the meta-analysis is limited by the strength of evidence reviewed which included case reports, case series, and observational studies, the results demonstrate propranolol is an effective treatment for infantile airway hemangioma.

Another meta-analysis published by Izadpanah et al. (2013) compared propranolol and corticosteroid use for treatment of hemangioma lesions. The analysis included 41 studies in total, involving 3424 subjects. Lesions were located on the trunk, extremities, head, neck and/or airway. A total of 2629 subjects received corticosteroids (oral or intralesional) and 795 received propranolol. Overall efficacy (regression of the lesion) for corticosteroid use was 69.1%, 17.6% developed side effects, rate of resolution was 84.5% receiving systemic and 66.4% receiving local administration. In comparison, the overall efficacy rate for propranolol was 97.3%, 13.7% developed side effects and rate of resolution was 98.9%. The response rate between propranolol and systemic corticosteroid use was statistically significant when intralesional studies were omitted, 97% versus 71% respectively. While the results of the meta-analysis are promising, the authors acknowledged it is limited by the lack of randomized controlled trials.

A systematic review published by Léauté-Labrèze and colleagues (2016) evaluating the safety of oral propranolol for treatment of infantile hemangioma provides support that treatment is well-tolerated if pretreatment assessments and within-treatment monitoring is performed. The systematic review included 5862 subjects who underwent treatment with oral propranolol and included manufacturers data (n=435 subjects), one Compassionate Use Program (n=1661 subjects), retrospective (n=44) and prospective (n=35) trials, noncontrolled (n=78) and nonrandomized (n=79) trials, as well as case series, cohort studies, and one open-label study versus historical control. The safety profile confirmed the use of oral propranolol for infantile hemangioma was similar to that observed when used for cardiologic indications. The authors report the most common related adverse events were transient, nonserious, and manageable; serious risks in certain cases can be avoided with appropriate screening and exclusion criteria, and in other cases it can be minimized and/or managed with appropriate monitoring, caregiver education, and discontinuation of therapy when necessary.

In a report from the Agency for Healthcare Research and Quality (Chinnadurai, et al., 2016) the authors systematically reviewed evidence addressing the diagnosis and management of infantile hemangiomas (IH). The authors summarized data qualitatively and quantitatively using meta-analysis and evaluated the strength of evidence. A total of 81 studies evaluated oral propranolol, oral nadolol, oral atenolol, or timolol. The authors concluded beta blockers demonstrated significantly greater effects on reducing lesion size versus placebo or observation (high strength of evidence) and was also superior over various comparators. High mean rates of IH clearance with oral propranolol (95%, 95% Bayesian credible interval [BCI]: 88%–99%) were reported using meta-analysis. Other beta blockers demonstrated promising effectiveness however the strength of the evidence was considered low.

Randomized controlled trials comparing oral propranolol to standard therapies for treatment of cutaneous hemangiomas are limited, nonetheless there is an increasing body of evidence to support clinical efficacy for regression of lesions and improved clinical outcomes. Based on the available evidence and acceptance in the medical community, and despite the need for further randomized controlled trials, evidence in the peer-reviewed published scientific literature supports clinical efficacy for oral propranolol as a treatment for infants with complicated hemangiomas. Published evidence evaluating safety and efficacy of other agents, used alone or in combination as treatment for infantile hemangioma, such as atenolol and topical timolol, has demonstrated promising results (Abarzua-Araya, et al., 2014; Ge, et al., 2015; Xu, et al., 2015; Puttgen, et al., 2016; Danarti, et al., 2016; Marey, et al., 2018). More recently, the AAP reported that topical timolol may be used to treat select thin and/or superficial IHs (B, moderate recommendation) (Krowchuk, et al., 2019).

Port Wine Stain

Port wine stains are a type of vascular malformation involving the superficial capillaries of the skin. They vary in size and location and are usually present at birth although not always clinically evident. In rare cases, a port wine stain may be referred to as “acquired” and become evident after injury to the skin or in association with hormonal influences (Legiehn, Heran, 2008). Most often, lesions are found on the face, neck, arms or legs. They may be related to other underlying conditions, such as Sturge-Weber syndrome. Sturge-Weber syndrome, also known as encephalotrigeminal angiomatosis, is characterized by a facial port wine stain in a trigeminal V1 (i.e., ophthalmic) distribution, leptomeningeal angiomatosis, and choroidal vascular malformation of the eye, which can lead to ipsilateral glaucoma and buphthalmos. Glaucoma occurs in 30% of patients with Sturge-Weber syndrome, and it develops before two years of age in 60% of these patients (Hussain, et al., 2004).

Port wine stains appear as sharply demarcated pink-red patches that darken with time and do not proliferate; growth of the lesion is dependent upon growth of the child. As the child matures, the lesion may become raised and exhibit red-to-purple nodules and papules in adult years, leading to potential disfigurement (e.g., pebbly and slightly thickened surfaces), and bleeding with trauma. Hypertrophy may develop in the soft tissue underlying the port wine stain. Early treatment may prevent the progression of development to hypertrophy and nodules in later years. It has been noted port wine stain lesions on the forehead or eyelids can be associated with ocular disorders and warrant frequent ophthalmology exams to prevent damage to the eye.

Laser devices such as the argon, carbon dioxide (CO₂), Nd:YAG, and copper vapor laser have been used to treat port wine stains. In many cases, these laser devices have been associated with poor cosmetic outcomes (Rothfleisch, et al., 2002). Pulsed dye laser therapy has been shown to be the most effective treatment for port wine stains; is associated with less adverse effects, including less post-operative scarring; and is considered the standard treatment of choice (Galbraith, et al., 2019; Tucci, et al., 2009; Yang, et al., 2005; Schmults, 2005). Evidence in the published medical literature suggests efficacy is increased if lesions are treated in infancy, although size and location are also predictors of outcome (Kelly, et al., 2019; Conlon, Drolet, 2004). Nonetheless, while most port wine stains lighten after a series of pulsed dye laser treatments, some cannot be completely removed (Kelly, et al., 2019; Yang, et al., 2005).

Other Vascular Lesions/Malformations

Vascular malformations are present at birth and typically grow as the child grows. There are various methods of describing vascular malformations. In general, simple malformations consist of one type of vessel (e.g., capillaries, lymphatic vessels, or veins), with the exception of arteriovenous malformations (AVMs) which consist of both arterial and venous. Combined malformations consist of two or more vessel abnormalities in any combination. Vascular malformations can also be described by flow, depending on the arterial component, as either slow flow or fast flow. Fast flow malformations may be associated with high-output heart failure (Ricci, et al., 2017).

Lymphatic (Lymphangioma): Lymphatic malformations consist of abnormally dilated lymphatic channels and commonly affect the head and neck area in children (lymphangioma). Most are present at birth although some may appear later in childhood as a result of infection or trauma. These lesions are either macrocystic or microcystic; microcystic are more difficult to treat and more often associated with complications. Aside from cosmetic concerns, depending on the size and location of the mass the lesion may be symptomatic. For example, when the oral and pharyngeal mucosa is involved there may be tongue swelling, tongue hypertrophy, mucosal bleeding, speech difficulty, and airway compromise. Common complications include disfigurement, infection and bleeding. Treatment is aimed at improving cosmesis, and alleviating any associated symptoms and involves surgical excision and/or sclerotherapy (Wetmore, Potsic, 2010; Tucci, et al, 2009; Morelli, 2011; Freiden, et al., 1997). Although used less frequently, other types of treatment such as scleroembolization and CO₂ laser have also been effective.

Arteriovenous: Arteriovenous malformations (AVM) of the skin are rare; however this type of lesion is a direct connection of artery to vein, bypassing the capillary bed. AVMs may appear at any time from birth to early adulthood and often remain stable for several years. They usually become noticeable at times of hormonal changes and at times may suddenly enlarge following infection or trauma (Tucci, et al., 2009). If the lesions are asymptomatic treatment is not necessary, however if ulceration and/or bleeding develop treatment is warranted and consists of embolization and excision (Wetmore, Potsic, 2010).

Venous: Venous malformations include but are not limited to vein only malformations and angiokeratomas. These lesions vary in size and may be superficial, deep or a combination of both. The lesions grow as the child grows but have a tendency to enlarge after direct trauma or with hormonal change such as during puberty or pregnancy from progressive ectasia of the vascular structure (Tucci, et al., 2009). For most lesions treatment is not necessary. When treatment is warranted, such as with pain from enlargement, treatment for superficial nodular lesions is surgical excision; larger deeper lesions may be treated with sclerotherapy. Other treatment modalities include ND:Yag laser therapy, endovenous laser therapy. In some instances treatments are combined to increase effectiveness however smaller localized lesions are usually managed with a single modality (Huang, Liang, 2010). Angiokeratomas are characterized by ectasia of the superficial dermal vessels with hyperkeratosis of the overlying dermal layer (Freiden, et al., 1997). They appear as flat hemangiomas with an irregular surface,

with surgical excision being the treatment of choice (Wetmore, Potsic, 2010). Although angiokeratomas are generally asymptomatic bleeding and itching may occur with trauma.

Fibro-Adipose Vascular Anomaly (FAVA): There is limited information in the peer-reviewed published scientific literature evaluating the occurrence of FAVA and treatment outcomes. FAVA has been described as a “new vascular entity”; reportedly a vascular-type malformation, occurring in children, teens or young adults, often associated with pain and dysfunction, occurring mainly in the extremities. The anomaly has been defined as “a fibro-adipose vascular anomaly involving veins that are engorged and intertwined with fibrofatty tissue in the muscle, and subcutaneous and cutaneous lymphatic malformation” (Shaikh, et al., 2016; Alomari, et al., 2017) and may be associated with contracture formation. It is a slow-flow vascular-type malformation (venous to lymphatic) (Alomari, et al., 2017). Conventional management generally includes observation, physical therapy, casts or splints, sclerotherapy, steroid injection, surgical excision, neurolysis or neurectomy, and Achilles cord lengthening (Shaikh, et al., 2016). Image-guided cryoablation has been effective in some children and young adults presenting with FAVA (Shaikh, et al, 2016). Authors generally agree however further studies are needed to better define the condition and effective clinical management.

U.S. Food and Drug Administration (FDA)

Lasers are regulated by the FDA as Class II devices and receive approval through the 510(k) process. According to the FDA, pulsed dye lasers are indicated for use in the treatment of cutaneous vascular lesions such as port wine stains and hemangiomas, and benign cutaneous lesions such as warts, striae and some forms of psoriasis.

Technology Assessment

Hayes, Inc. published a technology directory report evaluating the use of pulsed dye laser therapy for treatment of cutaneous vascular lesions (port wine stains and hemangiomas) (Hayes, 2012, reviewed 2016). According to the report, pulsed dye laser therapy was considered safe and effective for treatment of both port wine stain and hemangioma. Although definitive patient selection criteria has not been established the peer-reviewed published evidence supports the use of pulsed dye laser therapy in children and adults with port wine stains who require treatment to alleviate or prevent medical or psychological complications. The evidence was also sufficient to support efficacy for pulsed dye laser therapy as a treatment for superficial hemangiomas, the superficial component of mixed hemangiomas, post involutonal hemangiomas, and telangiectasia in infants or children requiring treatment to alleviate or prevent medical or psychological complications. Pulsed dye laser therapy is not efficacious for treatment of deep hemangiomas or for the deep component of mixed hemangiomas.

Professional Societies/Organizations

The American Academy of Pediatrics (AAP) published a clinical practice guideline for the management of hemangiomas (Krowchuk, et al., 2019). Within this guideline the AAP classifies IHs as high risk if there is evidence of or potential for life threatening complications, functional impairment, ulceration, structural anomalies or permanent disfigurement. The authors recommend early consult (by one month of age) for lesions that are potentially high risk and report that oral propranolol is the treatment of choice for problematic IHs that require systemic therapy.

Use Outside of the US: The National Institute for Health and Care Excellence (NICE) published a procedural guidance document regarding intralesional photocoagulation of subcutaneous congenital vascular disorders and noted that due to inadequate evidence supporting safety and efficacy the procedure should not be used without special arrangements for consent, audit or research (NICE, 2004). Guidance for laser and other modalities of treatment were not found.

Medicare Coverage Determinations

	Contractor	Policy Name/Number	Revision Effective Date
NCD	National	Laser Procedures (140.5)	5/1/1997
LCD		No Local Coverage Determination.	

Note: Please review the current Medicare Policy for the most up-to-date information.

Coding/Billing Information

- Note:** 1) This list of codes may not be all-inclusive.
 2) Deleted codes and codes which are not effective at the time the service is rendered may not be eligible for reimbursement.

Considered Medically Necessary when criteria in the applicable policy statements listed above are met:

Port Wine Stains

CPT®* Codes	Description
17106	Destruction of cutaneous vascular proliferative lesions (eg, laser technique); less than 10 sq cm
17107	Destruction of cutaneous vascular proliferative lesions (eg, laser technique); 10.0 to 50.0 sq cm
17108	Destruction of cutaneous vascular proliferative lesions (eg, laser technique); over 50.0 sq cm

Hemangiomas and Other Vascular Malformations

CPT®* Codes	Description
17106	Destruction of cutaneous vascular proliferative lesions (eg, laser technique); less than 10 sq cm
17107	Destruction of cutaneous vascular proliferative lesions (eg, laser technique); 10.0 to 50.0 sq cm
17108	Destruction of cutaneous vascular proliferative lesions (eg, laser technique); over 50.0 sq cm
37241	Vascular embolization or occlusion, inclusive of all radiological supervision and interpretation, intraprocedural roadmapping, and imaging guidance necessary to complete the intervention; venous, other than hemorrhage (eg, congenital or acquired venous malformations, venous and capillary hemangiomas, varices, varicoceles)
37242	Vascular embolization or occlusion, inclusive of all radiological supervision and interpretation, intraprocedural roadmapping, and imaging guidance necessary to complete the intervention; arterial, other than hemorrhage or tumor (eg, congenital or acquired arterial malformations, arteriovenous malformations, arteriovenous fistulas, aneurysms, pseudoaneurysms)
61626	Transcatheter permanent occlusion or embolization (eg, for tumor destruction, to achieve hemostasis, to occlude a vascular malformation), percutaneous, any method; non-central nervous system, head or neck (extracranial, brachiocephalic branch)

Oral Propranolol Administration to an Infant in the Inpatient Setting

HCPCS Codes	Description
J8499	Prescription drug, oral, nonchemotherapeutic, NOS

*Current Procedural Terminology (CPT®) ©2020 American Medical Association: Chicago, IL.

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